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Case reports

Aortic atresia with double inlet left ventricle, rudimentary left sided right ventricle, and ventriculoarterial discordance

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SUMMARY Aortic atresia associated with ventriculoarterial discordance (transposition) was found at necropsy in a 3 month old neonate. This is a rare association.

Aortic atresia is one of the most common causes of congestive heart failure and death in early life.[1] Usually, the great arteries are normally related. We report the unusual association of aortic atresia with ventriculoarterial discordance.

Case report

A 3 day old male neonate was referred with a history of tachypnoea and cyanosis during feeding. The blood pressure on admission was 75/40 mm Hg in the right arm and 90/50 mm Hg in the right leg. Physical examination detected a right ventricular impulse, single second heart sound, a grade 2/6 systolic ejection murmur along the left sternal border, an S_3 gallop, and hepatomegaly. A chest radiograph showed a cardiothoracic ratio of 0·65 with increased pulmonary vascularity. The electrocardiogram showed right atrial and left ventricular hypertrophy Arterial oxygen pressure was 51 mm Hg (6·8 kPa) in room air.

At cardiac catheterisation oximetry showed an increment in oxygen saturation at the right sided ventricular level with similar saturations in the pulmonary arteries and aorta. Pulmonary arterial pressure was equal to the systemic pressure. Ventriculography showed a large morphological left ventricle with ventriculoarterial discordance (Figure *ai* and *bi*). The catheter entered the aorta from the pulmonary artery via a duct. Aortography showed a left sided anteriorly positioned hypoplastic ascending aorta (Figure *aii* and *bii*).

The patient died in 1974 at 3 months of age of refractory congestive heart failure.

At necropsy visceroatrial situs solitus was present. Systemic venous drainage was into the morphological right atrium. The morphological left atrium received the four pulmonary veins. The normally formed atrial septum had a patent foramen ovale. The right sided atrioventricular valve had two leaflets and the left sided atrioventricular valve was tricuspid; both entered the left ventricle (a double inlet left ventricle). The origin of the posterior pulmonary artery was from this chamber. A rudimentary left sided right ventricle gave the origin to the anterior and leftward atretic aortic valve and hypoplastic ascending aorta. The great arteries were joined by a duct.

Discussion

Aortic atresia and ventriculoarterial discordance (transposition) are rarely found in tandem. To our knowledge only five such cases have been reported. Two were associated with double inlet left ventricles of the left ventricular type,[2 3] one with corrected transposition,[4] one with complete transposition,[5] and one with absent tricuspid valve with atrioventricular discordance.[6]

Systemic blood flow is rarely obstructed in hearts with ventriculoarterial discordance. A restrictive bul-

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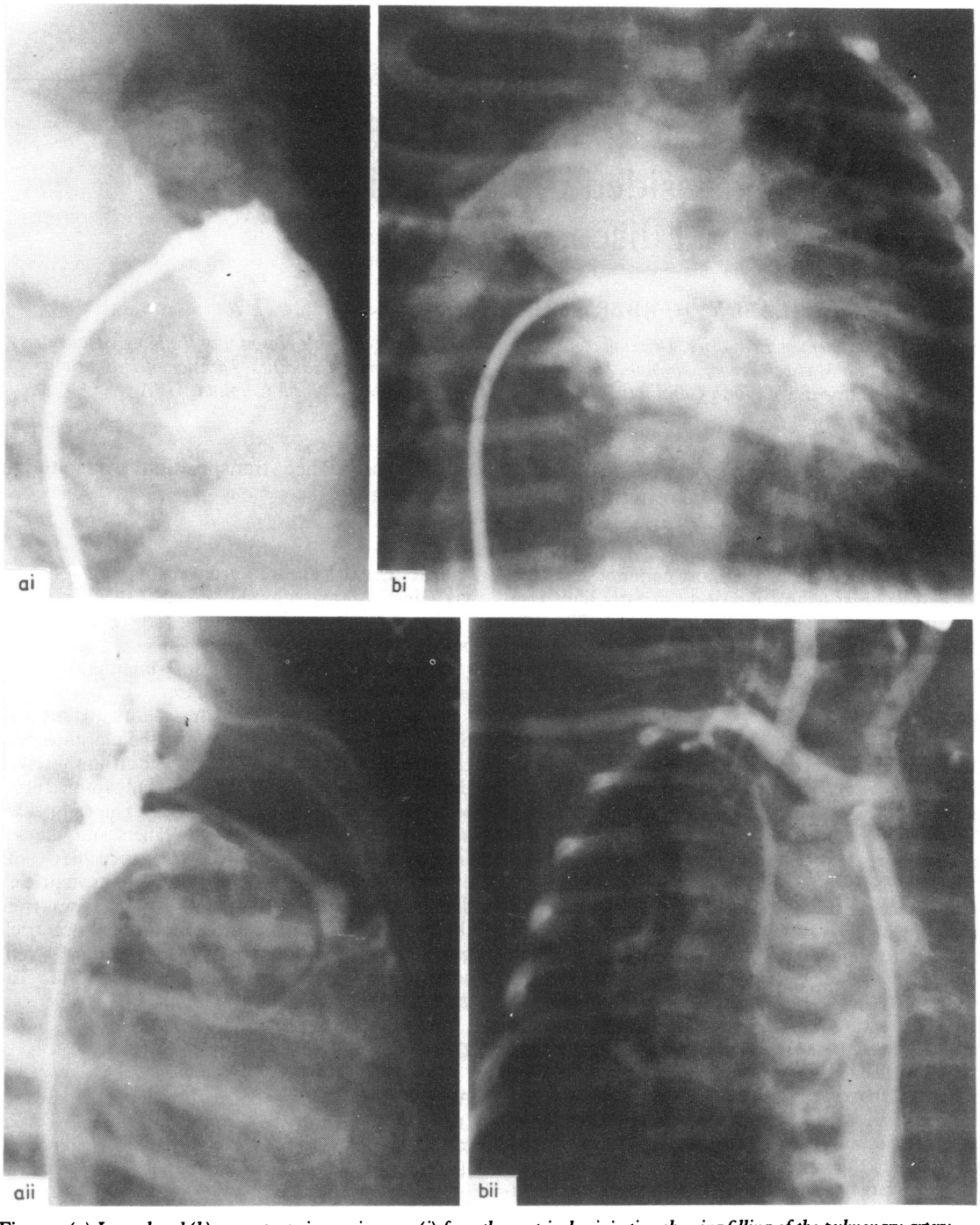

Figure *(a) Lateral and (b) anteroposterior angiograms (i) from the ventricular injection showing filling of the pulmonary artery, which is rightward and posterior in position (no contrast exits into the aorta from this injection), and (ii) from the retrograde aortic injection showing filling of the pulmonary artery which is posterior to the hypoplastic ascending aorta; the aorta is leftward and anterior in relation to the pulmonary artery.*

boventricular foramen and accessory endocardial cushion tissue are among the pathological mechanisms that limit aortic flow by producing subaortic stenosis or atresia.[7] Why this occurs is a matter of speculation.

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